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Radiology in Focus

Monostotic fibrous dysplasia of the temporal bone

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Abstract
Fibrous dysplasia is a slowly progressive bony disorder where normal bone is replaced by abnormal fibro-osseous tissue. Its monostotic variety in the temporal bone is very rare and such a case is presented here. Computed tomography (CT) may be adequate for the diagnosis and follow-up of these patients. Limited surgery should only be considered in cases of symptomatic disease.

Key words: Fibrous dysplasia, monostotic; Temporal bone

Introduction

Fibrous dysplasia is a slowly progressive, expansile bony dysplasia of unknown aetiology in which normal bone is replaced by abnormal fibro-osseous tissue. It may be monostotic or polystotic. Craniofacial involvement is found in only 10 per cent of cases of the monostotic variety, while monostotic fibrous dysplasia of the temporal bone is very rare (Brown *et al.*, 1995).

Case report

A 47-year-old man presented with several months history of progressive hearing loss and tinnitus in the left ear. There were no other ear-related symptoms and his medical history was unremarkable. Otoscopy revealed a narrow left external ear canal and through it part of a normal looking tympanic membrane could be seen. Rinne was negative on the left side and Weber lateralized to the left. On clinical examination, a hard non-tender swelling was felt over the left pre- and post-auricular regions. The rest of the examination was normal. Pure tone audiometry demonstrated a 40 dB air-bone gap with a Caarharts notch at 2000 Hz. Impedance audiometry gave a type A tympanogram. A CT scan was obtained (Figure 1) which showed sclerosis and marked focal expansion of the left squamous temporal bone, with a 'ground glass' appearance, which is characteristic of fibrous dysplasia. A T1-weighted magnetic resonance image (MRI) scan with gadolinium DTPA revealed expansion of the squamous temporal bone with loss of normal architecture of the cranial vault and diffuse low signal due to sclerosis. There is an enhancing extradural component medial to the osseous mass, which indents the temporal lobe. This may represent a fibrous component. The left internal auditory meatus and the right temporal bone were normal (Figure 2). Biopsy of the lesion confirmed fibrous dysplasia of the temporal bone.

Discussion

Fibrous dysplasia is a relatively uncommon bone disease in which normal bone is replaced by abnormal fibro-connective tissue proliferation. Although its aetiology is

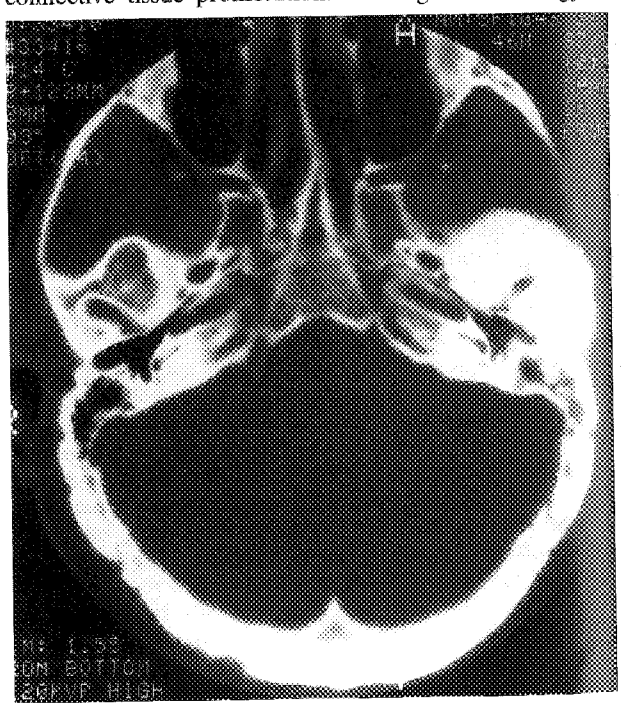

FIG. 1

Fibrous dysplasia of the left temporal bone. Axial CT shows focal expansion of the squamous temporal bone and petrous base with sclerosis and a 'ground glass' appearance. Anteriorly the abnormality is limited by the sphenotemporal suture. There is minor bone encroachment on the attic, but not on the labyrinth or internal auditory canal.

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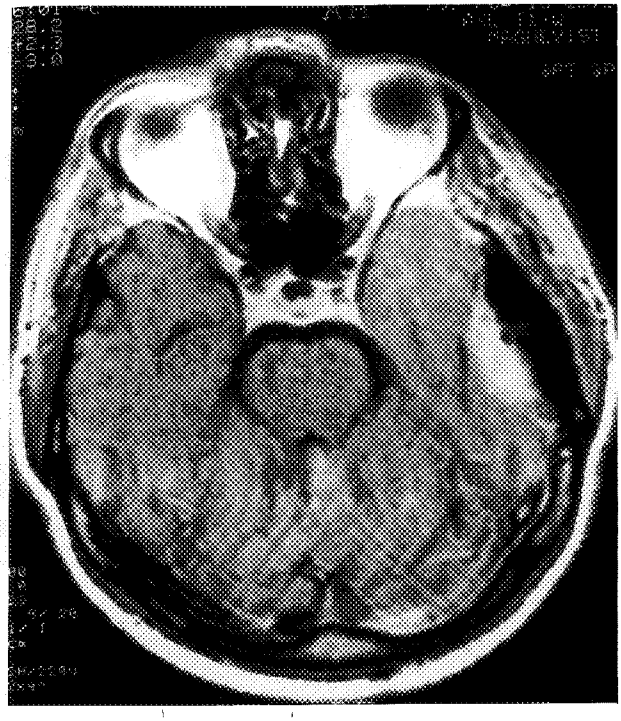

FIG. 2

MRI scan of the same patient. A post-gadolinium T1-weighted image revealed thickening of the squamous temporal bone, which is of homogenous low signal, due to sclerosis and loss of differentiation of the inner and outer tables of skull and diploic space. There is a broad band of enhancing tissue medial to the expanded bone that represents a non-osseous component, probably fibrous.

still unknown, it is considered by some authors to represent a hamartomatous malformation resulting from an idiopathic arrest in the maturation of bone at the woven bone stage (Mohamadi-Araghi and Haery, 1993). Histologically fibrous dysplasia is heterogenous with regions of predominantly soft tissue or bone. Soft areas are abundant in collagen and are mostly acellular. The bony areas have trabeculae of immature woven rather than lamellar bone (Nager *et al.*, 1982; Cheung and Jackler, 1994).

The disease may affect one bone (monostotic) or several bones (polystotic). The McCune-Albright syndrome is characterized by polystotic fibrous dysplasia with associated endocrinopathy and skin hyperpigmentation. In the monostotic form involvement of the temporal bone is very rare with fewer than 60 cases reported in the literature.

The most common clinical manifestations of temporal bone fibrous dysplasia include conductive hearing loss, external ear canal stenosis, and swelling of the temporal bone (presenting usually in the post-auricular area). Cholesteatoma can form as a result of entrapped squamous epithelia deep to an obstructed external auditory meatus. Sensorineural hearing loss is rare and may develop secondary to a labyrinthine fistula in the presence of cholesteatoma or as a result of internal auditory meatus stenosis. The otic capsule is spared by the disease (Cheung and Jackler, 1994). Facial nerve paralysis is rare but can develop secondary to chronic suppurative otitis media or fallopian canal stenosis (Nager *et al.*, 1982). Sarcomatous transformation has not yet been reported in the temporal bone, but it has been in other sites.

Radiologically three different types have been classified: the pagetoid (56 per cent), the radiolucent (21 per cent) and the sclerotic type (23 per cent). Solitary lesions on plain radiographs should be distinguished from non-ossifying and ossifying fibroma, simple bone cyst, eosinophilic granuloma, giant cell tumour, osteoblastoma, Paget's disease, and osteogenesis imperfecta. Intradiploic meningioma is an important differential diagnosis in the skull base. High-resolution CT findings may be pathognomonic and offer a great advantage in diagnosing as well as monitoring the progress of the disease (Reddy *et al.*, 1994). CT is useful in assessing the degree of external auditory canal stenosis, involvement of the middle-ear cleft and internal auditory meatus. CT characteristics of fibrous dysplasia include expansion of the involved bone with either heterogenous density (sometimes with scattered islands of bone formation) or a homogenous 'ground glass' appearance, as also seen on plain radiographs. Involvement of the optic canals, superior and inferior orbital fissures, frontal recess and osteomeatal complex can best be evaluated by CT scan (Mohammadi-Araghi and Haery, 1993). On MRI fibrous dysplasia usually exhibits a low to intermediate signal on T1-weighted images and heterogenous low signal on T2-weighted images. After intravenous gadolinium-DTPA (Gd-DTPA) the lesions usually show a moderate to marked enhancement (Casselman *et al.*, 1993; Mohammadi-Araghi and Haery, 1993). Signal intensity on T1- and T2-weighted images and the degree of enhancement following Gd-DTPA depends on the relative proportions of bony trabeculae, cellularity, collagen, and cystic changes.

In our case there was clinical evidence of increase in the size of the temporal bone presenting as a hard pre- and post-auricular swelling. As the MRI scan revealed, the lesion expanded into the middle cranial fossa. The enhancing extradural component of the mass causes marked indentation of the temporal lobe. This component was not identified on the previous contrast-enhanced CT scan. Although there is no such history, this lesion may be potentially epileptogenic. Association of fibrous dysplasia of the temporal bone with epilepsy has been reported in the literature (Nishioka, 1982).

There is no specific medical treatment for fibrous dysplasia. Asymptomatic monostotic lesions can be monitored regularly without any intervention. Surgical management is justified if the lesions becomes symptomatic and includes maintenance of a wide external auditory meatus to stabilize hearing, and management of secondary complications arising from entrapped cholesteatoma that may interfere with cochlear, vestibular or facial nerve function. The surgeon should be aware that surgery of the dysplastic temporal bone can be hazardous because landmarks are often obliterated and intra-operative bleeding can be vigorous (Cheung and Jackler, 1994). Restenosis of the external ear canal can occur. This may be prevented by the use of a permanent silastic stent. There is now evidence that in a proportion of affected adults, fibrous dysplasia is an active process (Davies and Macpherson, 1991) and as such it should be followed up clinically and radiologically and should not be treated as an incidental finding.

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